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# Myelodysplastic syndromes

The incidence of primary myelodysplastic syndromes (MDS) is about 5 per 100,000 persons per year in the general population, but it increases to 20-50 per 100,000 persons per year after 60 years of age. About 25,000 new cases are therefore expected in the Europe each year. The FAB classification has been widely used for defining the different subtypes of MDS. More recently, the World Health Organization (WHO) classification of myeloid neoplasms has proved to be a very useful tool. The WHO classification provides valuable prognostic information and may be used for clinical decision making [Malcovati *et al.*, J Clin Oncol 2005;23:7594-603]. The International Prognostic Scoring System (IPSS)3 – based on the percentage of marrow blasts, the cytogenetic pattern, and the number and degree of cytopenias – is useful for predicting survival and the risk of leukemia. We found that dependency on transfusions has a negative effect on the likelihood of survival [Cazzola & Malcovati, N Engl J Med 2005;352:536-8]. Based on WHO subgroups, karyotype, and transfusion requirement, we defined a time-dependent prognostic scoring system (WPSS) that can be used to predict survival and leukemia progression at any time during the clinical course of the disease. More recently, we showed that bone marrow fibrosis represents an additional independent prognostic factor, as this feature identifies a distinct clinical entity characterized by high transfusion need and poor prognosis. In addition, the presence of non-hematologic comorbidities significantly worsens the survival of MDS patients, and this must be taken into account in risk assessment. A risk-adapted treatment strategy is mandatory for disorders that range from indolent conditions lasting years to forms approaching acute myeloid leukemia. At present, the only treatment that can definitely prolong survival is allogeneic hematopoietic stem-cell transplantation. However, less than 10 percent of all MDS patients are eligible for such treatment and have a donor. Three drugs have been approved by the US Food and Drug Administration (FDA) for the treatment of MDS. Azacitidine has been approved for treatment of patients with the following MDS subtypes: refractory anemia or refractory anemia with ringed sideroblasts (if accompanied by neutropenia or thrombocytopenia or requiring transfusions), refractory anemia with excess blasts, refractory anemia with excess blasts in transformation, and chronic myelomonocytic leukemia. Lenalidomide has been approved for treatment of patients with transfusion-depen-